

CASE REPORT

A 72 year old woman with ALCAPA

C Fierens, W Budts, B Denef, F Van de Werf

Abstract

ALCAPA syndrome (anomalous origin of the left coronary artery from the pulmonary artery), which causes the left coronary artery to grow with an anomalous origin from the pulmonary artery, is a rare disease which may result in myocardial infarction, congestive heart failure, and sometimes death during the early infantile period. A 72 year old woman with ALCAPA syndrome is presented. The asymptomatic patient presented with a cardiac murmur which was discovered during a routine check up for a gynaecological intervention. Coronary cineangiography established the diagnosis. Although surgical correction is the usual treatment for such cases, medical treatment was preferred for this patient because she was asymptomatic without clinical signs of heart failure.

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Department of
Cardiology, University
Hospital Gasthuisberg,
Herestraat 49, B-3000
Leuven, Belgium

C Fierens
W Budts
B Denef
F Van de Werf

Correspondence to:
Dr Budts

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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiovascular defect.¹⁻⁶ An extended left ventricle infarction can be fatal if the defect is not corrected surgically in the first years of life.²⁻⁵⁻⁹ Patients with ALCAPA syndrome who survive past childhood often have varying symptoms of myocardial ischaemia or heart failure, depending on the development of collateral circulation, and up to 90% die suddenly at a mean age of 35 years.²⁻⁶ There are few patients older than 50 who have ALCAPA syndrome that has not been surgically treated.

Until now only 26 patients have been described.⁴ We report a 72 year old woman with ALCAPA, who was almost asymptomatic in spite of a significant left to right shunt.

Case report

A 72 year old woman was referred to our hospital to determine the clinical importance of a continuous heart murmur which had been discovered during a routine check up before a gynaecological procedure. She had no cardiac antecedents but at presentation she mentioned mild shortness of breath (New York Heart Association functional class I) and palpitations during heavy exercise.

Physical examination revealed a continuous murmur in the 3rd and 4th parasternal intercostal spaces (grade 3/6) and a mild holosystolic murmur at the apex. There were no clinical signs of left or right heart failure. ECG showed lateral repolarisation abnormalities (fig 1). Chest x ray revealed cardiomegaly and prominent vascular trauma (fig 2), and trans-thoracic echocardiography showed a normal right ventricle and a mildly dilated left ventricle with a minimal hypertrophic and hypokinetic interventricular septum. Left ventricular ejection fraction, measured in M mode, was 74%. No septal defects were found and colour Doppler imaging revealed only mild mitral and tricuspid regurgitation. Estimated systolic right ventricular pressure was 40 mm Hg, and we observed a dilated blood vessel with an

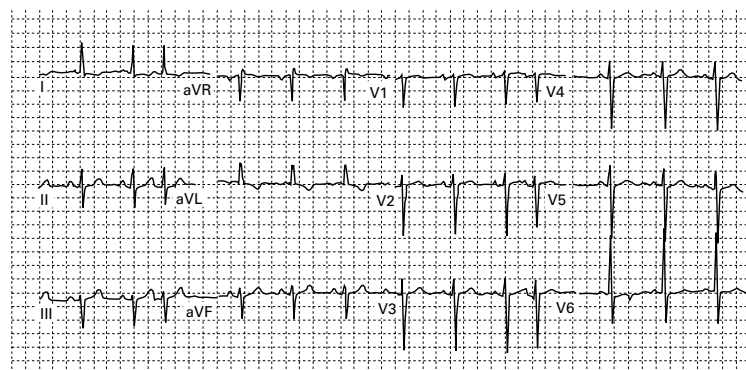


Figure 1 Electrocardiogram showing lateral repolarisation abnormalities.



Figure 2 Chest x ray showing cardiomegaly and prominent vascular trauma.

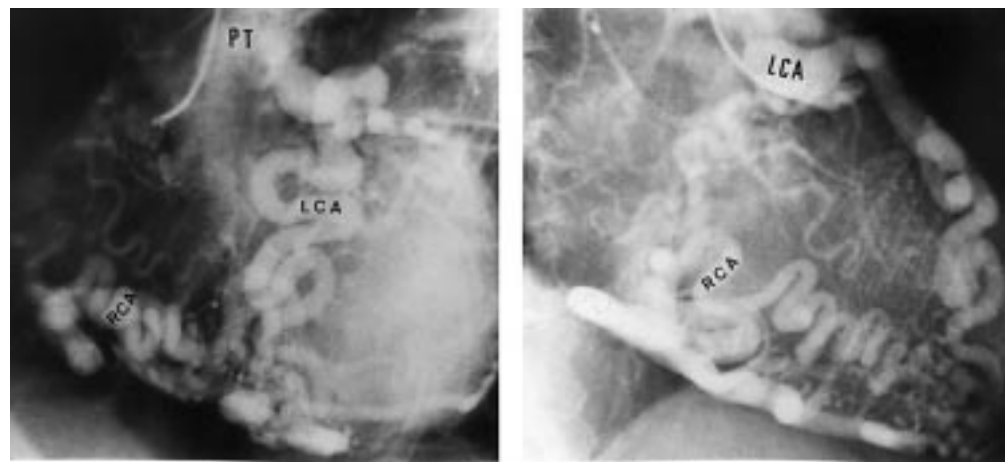


Figure 3 Coronary angiography showing an extremely dilated and tortuous right coronary artery (RCA) with strong collateral filling of the left coronary artery (LCA) shunting into the pulmonary trunk (PT).

approximate diameter of 1 cm. It seemed to originate from the ventral side of the aorta, with prominent laminar diastolic flow compatible with coronary artery flow. A referring cardiologist suggested a single coronary artery or an important coronary fistula.

A transoesophageal echocardiogram showed a severely dilated right coronary artery originating from the right coronary cusp and connecting distally to the pulmonary trunk 2 cm above the pulmonary valve. The origin of the left coronary artery in the left coronary cusp could not be found.

The diagnosis of ALCAPA was made by coronary angiography, which revealed an extremely dilated and tortuous right coronary artery with strong collateral filling of the left coronary artery, shunting into the pulmonary trunk (fig 3). Saturation sampling during right heart catheterisation showed a mild left to right

shunt ($Q_p/Q_s = 1.5/1$) with a clear saturation gap of 10% between the lower and the higher section of the pulmonary trunk (table 1). The suspected pulmonary hypertension which had been indicated by echocardiography was confirmed (table 2). Nuclear magnetic resonance was also able to locate the origin of this vessel (fig 4).

Conservative treatment with an angiotensin converting enzyme inhibitor was proposed, due to the age of the patient and the fact that she

Table 1 Saturation sampling during right heart catheterisation showing a mild left to right shunt ($Q_p/Q_s = 1.5/1$) with a clear saturation gap of 10% between the lower and the higher section of the pulmonary trunk

	Oxygen saturation (%)
Vena cava inferior	65.6
Vena cava superior	62.1
Right atrium high	61.5
Right atrium mid	63.1
Right atrium low	53.3
Right ventricle high	62.2
Right ventricle mid	61.6
Right ventricle low	59.2
Arteria pulmonalis trunk low	68.7
Arteria pulmonalis trunk high	78.3
Arteria pulmonalis right	71.8
Arteria pulmonalis left	73.8
Arterial	95.9

Table 2 Pressures measured during right heart catheterisation showing pulmonary hypertension (mean 30 mm Hg)

Pressure	Systolic (mm Hg)	Diastolic (mm Hg)	Mean (mm Hg)
Right atrium	—	—	7
Right ventricle	40	9	—
Arteria pulmonalis	40	20	30
Pulmonary capillary wedge pressure	23	—	—
Aorta	170	84	—
Left ventricle	176	27	—

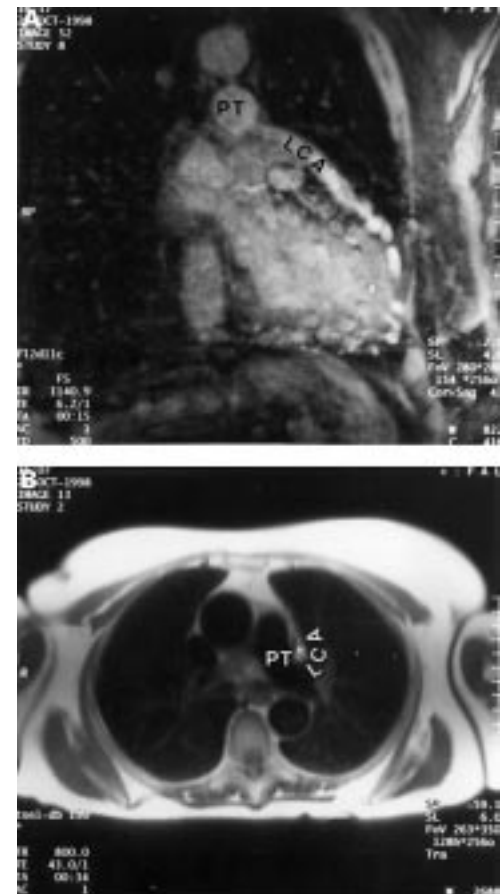


Figure 4 Nuclear magnetic resonance scan showing a dilated left coronary artery (LCA) merging with the pulmonary trunk (PT). (A) T1 weighted longitudinal section; (B) T2 weighted cross section.

was asymptomatic without clinical signs of heart failure.

Discussion

ALCAPA was first mentioned in 1908, in a 60 year old woman.² The syndrome was also described by Bland *et al* in 1933 after the study of the clinical and necropsy findings from an infant with symptoms of dyspnoea and myocardial infarction.¹

The estimated incidence of ALCAPA is 1/300 000 live births (between 0.24% and 0.46% of all congenital cardiac anomalies).^{3–10} Some believe that this is a significant underestimation of the true incidence, as many patients may be asymptomatic until their death and therefore remain undiagnosed.¹⁰ This case confirms the possibility that many cases may remain undetected.

ALCAPA develops before birth when the systemic and pulmonary arterial pressures are equal and there is antegrade flow in both the left and the right coronary. In the neonatal period this gradually changes as the pulmonary blood pressure diminishes, the ductus arteriosus closes, and the flow in the left coronary artery reverses.⁶ The development of collateral circulation between the right and left coronary artery during closure of the duct and lowering of the pulmonary pressure will determine the extent of myocardial ischaemia.⁷

If not treated surgically, up to 85% of patients with ALCAPA die in infancy or childhood,^{2–8} and untreated ALCAPA in the elderly is extremely rare. In 1995, Alexi-Meskishvili and colleagues found only 26 patients over 50 years old.⁴ The clinical presentation of older ALCAPA patients is usually non-specific: syncope, cardiac arrhythmias, exertion fatigue, nocturnal dyspnoea and less commonly, angina pectoris.² The defect is sometimes discovered during necropsy after sudden death.⁴

Clinical examination reveals a systolic murmur in almost 70% of cases.^{2–10} As in our patient, the ECG usually shows lateral ischaemia or sequels of a (anterior) myocardial infarction.² Bicycle testing, myocardial scinti-

graphy, positive emission tomography, and dobutamine stress echocardiography can all reveal chronic ischaemia, which can be improved through surgery.^{4–8} Diagnosis is usually established by a coronary angiogram, which shows a dilated and tortuous right coronary artery with collateral filling of the left coronary artery system; variable degrees of shunting to the pulmonary artery may be present. Right heart catheterisation may show an increase in oxygen saturation at the point where the blood of the left coronary artery merges with the blood from the pulmonary artery.² Surgical treatment usually recreates a dual coronary perfusion, regardless of the presence of symptoms or state of myocardial function.^{4–10} However, in our patient, who may be the oldest to be reported with this anomaly,⁴ the risk of cardiac surgery seemed to outweigh the natural prognosis. Therefore we chose not to intervene surgically.

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